



ADPKD in children

Dr Heather Lambert

Consultant Paediatric Nephrologist

Beware names

- Polycystic kidney disease
 - Adult = autosomal dominant ADPKD
 - Infantile = autosomal recessive ARPKD
- Multicystic kidney disease
- Others eg Cystic dysplastic

ADPKD in childhood

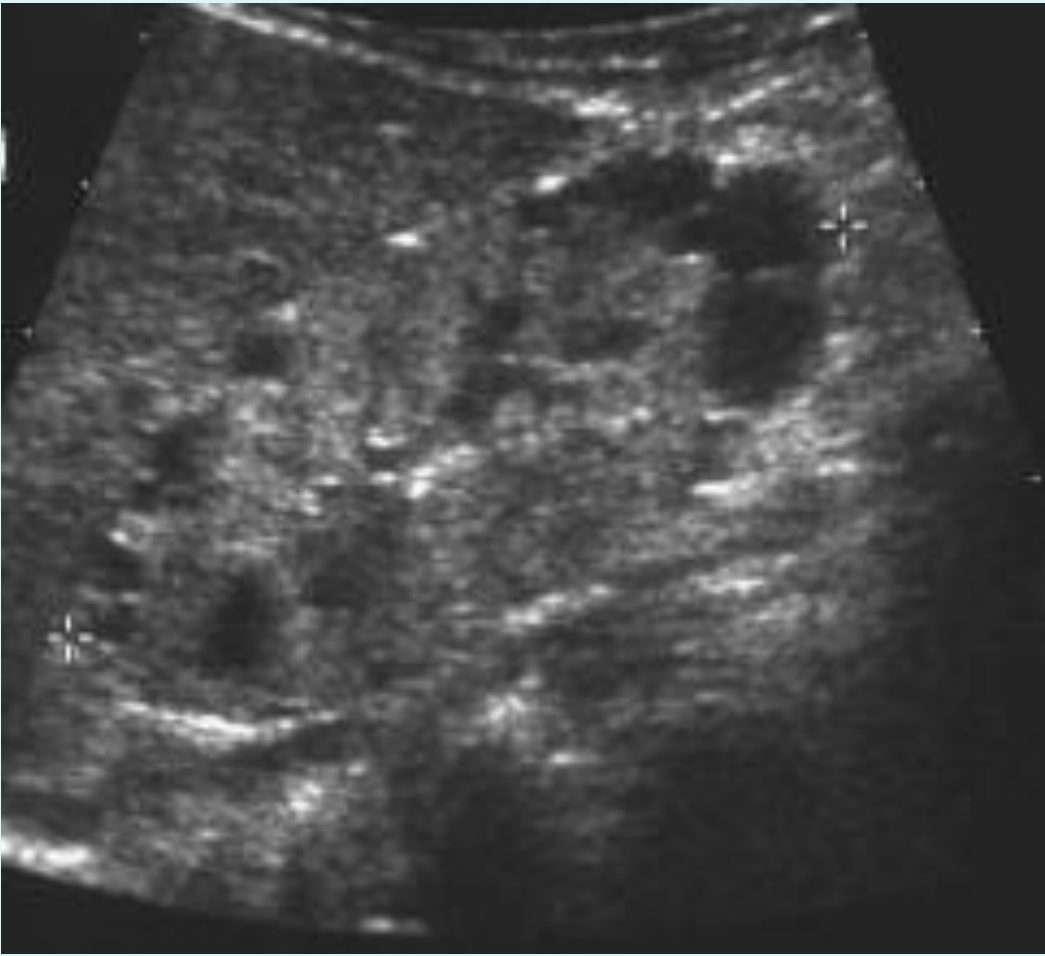
4 ways

- On routine antenatal US scan
- Found on kidney US scan
 - eg scan done for tummy ache
 - or scan done after urine infection
- Found to have hi BP
- Screening

Antenatal detection- diagnosis often unclear



- Take full family history
- Offer to scan both parents
- Arrange postnatal scan, bloods, urine & BP on baby



May present with related problem

- Urine infection
- High BP
- Blood in urine
- Tummy ache or loin/ back pain

Screening

Referrals often from medical obstetric clinic

Offer

- Yearly BP and urine test
- With or without US scan (in 1st year & 5yearly)

Parents choice

(but I strongly recommend BP checks)

Management

- Pick up hi BP before causes problems/damage
- Treat BP really well
- Treat protein in urine
- Advise avoid ibuprofen (esp if bit dehydrated)
- Drink well & avoid dehydration (may not concentrate urine well)
- Treat urine infections promptly
- Rarely have non-renal ADPKD problems in childhood

- Is the outcome more severe if you present as a young child?

- If one child in a family presents early will affected brothers & sisters also present early?

- How can a baby be found to have polycystic kidneys when neither parent has it on scan?